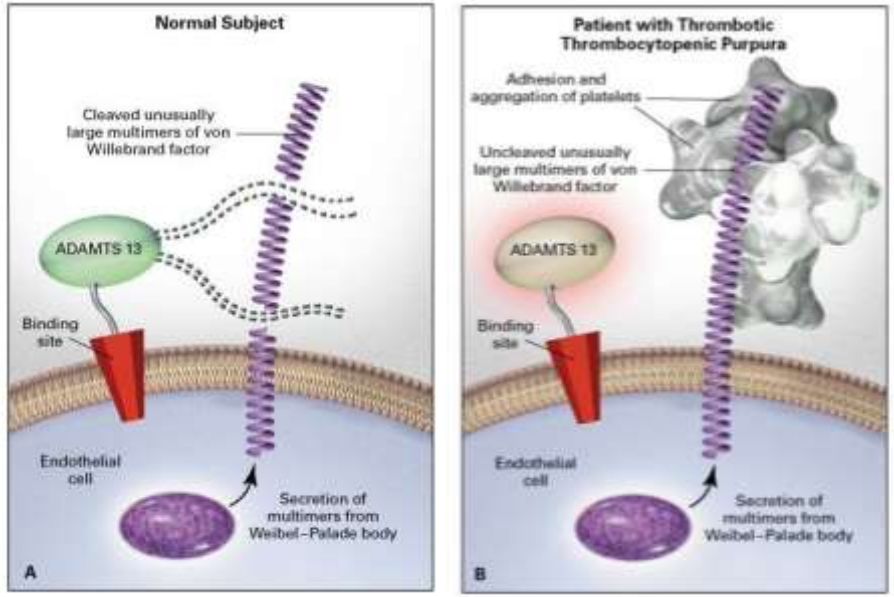
THROMBOTIC THROMBOCYTOPENIC PURPURA

(TTP)

Introduction



- **Autoimmune** disorder
- **Rare** disorder, with annual incidence – 1/75,000
- Mediated by **Antibodies against** metalloproteinase enzyme, **ADAMTS-13**
- Normally, ADAMTS-13 cleaves von Willebrand Factor (vWF) multimers into normal functional units (ADAMTS-13 also called **von Willebrand factor-cleaving protease (VWFCP)**

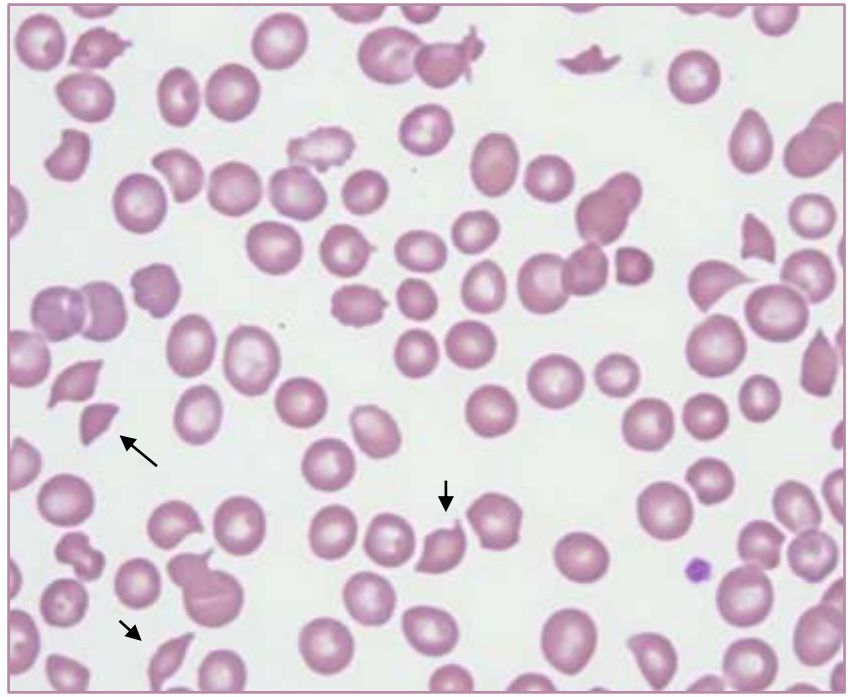
Deficiency of ADAMT-13 → **Large vWF multimers** → **Cross-link platelets** → **Thrombosis & Throbocytopenia**

Features

- Depends upon microvascular occlusion by platelet thrombi of principal organs, mainly brain and kidneys
- **Pentad** of findings, including:
  1. Thrombocytopenia
  2. Microangiopathic haemolytic anaemia (MAHA)
  3. Neurological sequelae
  4. Fever, &
  5. Renal impairment
- TTP may occur alone, or in association with Drugs (Ticlopidine, Ciclosporin), HIV, Shiga toxins, Pregnancy & Malignancy

Investigations

- **CBC** – Thrombocytopenia & Anemia
- **Renal Function Tests** - Impaired
- *Elevated* unconjugated bilirubin, LDH, reticulocytes, & *Low* haptoglobin
- **Peripheral blood film microscopy** - Features of MAHA like schistocytes & helmet cells
- **ADAMTS-13 activity** - <10% activity level
    - Useful in distinguishing TTP from HUS
    - Early therapy with plasma exchange is crucial in TTP



Treatment

- Emergent **Plasma Exchange**
- **Glucocorticoids, Aspirin & Rituximab** – also have a role
- **Platelet transfusion is contraindicated**

Prognosis

- Untreated, mortality rates are 90% in the first 10 days
- Even with appropriate therapy, the mortality rate is 20–30% at 6 months

Last Second **Medicine**

Like . Share . Subscribe